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ACUTE FEBRILE NEUTROPHILIC DERMATOSIS (SWEET SYNDROME) - A RARE CASE REPORT WITH LITERATURE REVIEW

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ABSTRACT

Introduction: Sweet syndrome is a rare skin disorder of unknown etiology. It is characterized by fever and the sudden onset of a rash. The typical age of onset is between 30 and 60 years of age, although cases have been reported in the paediatric and elderly populations as well. **Case presentation:** This study presents a case of an 8 year old male patient who had a seven day history of fever, lesions over the skin and redness in both eyes. She also presented with mouth ulcers in the last 4 days. Cultures, a biopsy and laboratory tests yielded a diagnosis of sweet syndrome. The case was managed successfully by pharmacotherapy and was resolved within 2 months. **Discussion**: Sweet syndrome is a very rare inflammatory skin disorder in children and has only 80 reported cases in the medical literature. Diagnosis primarily depends on histopathological examination. Treatment with corticosteroids is considered the ideal choice for such pathology. **Conclusion:** This case is exceptional because the occurrence of sweet syndrome in children is extremely rare and accurate diagnosis is necessary to initiate effective treatment. Its head, neck and oral manifestations and its association with paraneoplastic disease warrant the need to be aware of the condition.

KEYWORDS: paraneoplastic, autoimmune, idiopathic, bullous pyoderma, gangrenosum.

INTRODUCTION

Sweet syndrome was originally described by Dr. Robert Douglas Sweet and published in the British Journal of Dermatology as an 'Acute febrile neutrophilic dermatosis. The distribution of Sweet syndrome cases is worldwide and there is no racial predilection.

Sweet syndrome has been associated with autoimmune processes, malignancies, infections, drug reactions and gastrointestinal disorders such as inflammatory bowel disease. Based on this, it can present in several clinical settings as: classical (or idiopathic) Sweet syndrome, malignancy-associated Sweet syndrome and drug induced Sweet syndrome.

Affected patients present with an abrupt onset of tender plaques or nodules and accompanying fever, arthralgias, ophthalmologic manifestations, headaches and in rare cases, oral and genital lesions. Some authors suggest that the mechanism of action involves a dysregulation of immune function or an immunomediated hypersensitivity.

The diagnosis of Sweet syndrome is confirmed by biopsy. The findings are significant for a neutrophilic dermatosis with interface involvement of the epidermal/dermal junction. In this article, a rare case of Sweet syndrome with oral manifestations is described.

CASE REPORT

An 8-year-old male patient presented to an otolaryngologist (R.B.C.) with severe pain in his tongue and throat and referred pain in his right ear, along with odynophagia and hoarseness of 48 hours' duration. He also reported a fever, insidious in onset, associated with chills and rigors, burning micturition and increasing fatigue and malaise. He presented with redness in the right eye which was insidious in onset, associated with pain and tearing. He denied genital lesions and urethral discharge.

An oral and oropharyngeal examination revealed the presence of aphthous ulcerations, as well as a 3 × 3-cm raised inflammatory lesion on the right anterior lateral tongue and a 5 × 5-mm bulla on the hard palate in the midline. Also noted was warm, tender swelling of the left ankle, hand, and wrist. Because of the pain and swelling of the left hand and wrist, the patient was unable to completely close his hand. Erythematous papules and macules were noted on his face, neck, thighs and extremities. Nasolaryngopharyngoscopy detected diffuse inflammatory changes, but no specific lesion or disease process was identifiable.

The patient was initially started on an analgesic and oral cetirizine 10 mg/day. He was then referred to a dermatologist. By the time of the dermatology consultation, the raised lesion on the right lateral tongue had progressed to a 2.5-cm erosion. Fissuring was also seen in the right corner of the patient's mouth. Minimal gingival erythema was present. Scleral involvement was noted. On the upper and lower extremities, erythematous papules and plaques were present; painful, intact, bullae were also present on the lower extremities (figure 1 and 2). No involvement of the trunk, genitals, palms, or soles was present.

The working differential diagnosis included bullous erythema multiforme, bullous staphylococcal infection, pemphigus vulgaris, and Sweet syndrome. The bullae were subjected to bacterial culture, and the tongue ulcer was subjected to viral culture. Punch biopsy of an intact bulla on the right thigh was performed, and blood was drawn for evaluation. The patient was started IV Dexamethasone 8 mg, IV pheniramine 20 mg and for the pain he was given oral diclofenac at 50 mg twice daily. For the redness in the eye he was prescribed with ketorolac eye drops 0.5% TID. To relieve the moth ulcers, topical choline salicylate of 9%w/v TID was prescribed.

The biopsy results revealed both a superficial and deep perivascular and interstitial infiltrate composed mostly of neutrophils (figure 3 and 4). Eosinophils were also present. These findings were consistent with Sweet syndrome. Laboratory testing revealed that the white blood cell count was slightly elevated at12,500 cells/µl (normal: 4,000 to 11,000). The platelet count was elevated at 470 × 103 µl (normal: 140 to 400), as was the neutrophil count at 9,833 cells/µl (normal: 1,500 to 7,800). Both the hemoglobin and hematocrit values were decreased at 11.2 g/dl and 31.1%, respectively (normal: 13.2 to 17.1 and 38.5 to 50%, respectively). The viral culture was negative, but the bacterial culture revealed growth of group B streptococci and Staphylococcus aureus.

The patient was then prescribed with oral prednisone at 60 mg/day, with a slow taper over a 2-month period, oral trypsin-chymotrypsin TID and oral paracetamol 500mg TID for 14 days following which his oral and cutaneous lesions resolved. He was advised to undergo a thorough paraneoplastic workup, and he was made aware of the possibility of developing other diseases associated with Sweet syndrome, including gastrointestinal disorders, malignancy, and leukaemia.

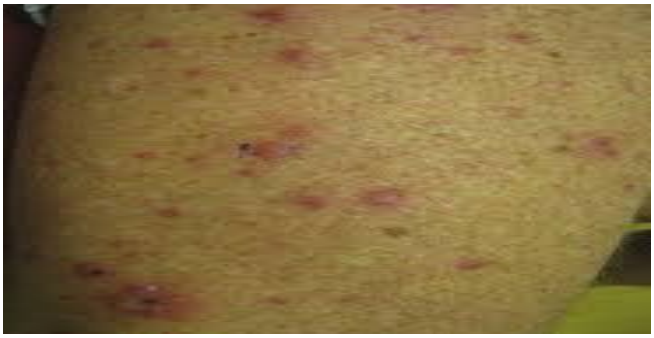

Figure 1. The right leg exhibits erythematous papules and tense, intact bullae.

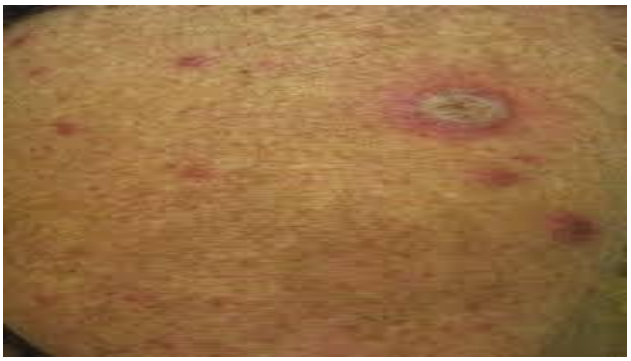

Figure 2. Erythematous papules and plaques are seen on the left leg.

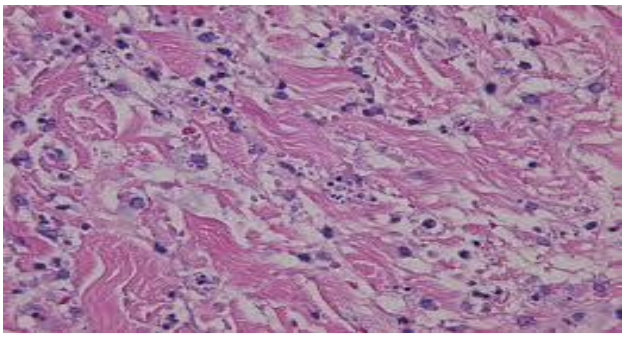

Figure 3. Histopathology showing the superficial and deep perivascular and interstitial infiltrate at low power.

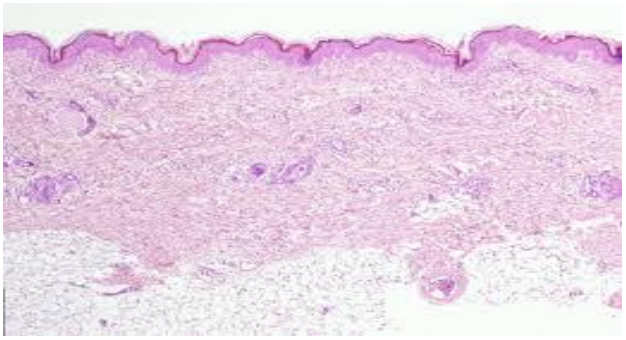

Figure 4. Histopathology showing the superficial and deep perivascular and interstitial infiltrate at high power.

DISCUSSION

Sweet syndrome has no racial predilection; It is reported to be more common in females than males by a ratio of 4:1. Ages of affected patients have ranged from 2 to 75 years; however most cases have occurred between the ages of 30 and 50 years.[1] Although its etiology is unknown, Sweet syndrome has been associated with many conditions. Sweet syndrome can present in several clinical settings as: classical or idiopathic (most common), malignancy-associated, and drug-induced sweet syndrome. Classical or idiopathic Sweet syndrome predominantly affects women and may be associated with infection (upper respiratory tract or gastrointestinal tract), inflammatory bowel disease or pregnancy.[2,3] Malignancy-associated Sweets syndrome is most often associated with acute myelogenous leukaemia. Carcinomas of the genitourinary organs, breast and gastrointestinal tract are the most frequently occurring cancers in Sweets syndrome with dermatosis-related solid tumors.[4,5]

Drug-induced Sweet syndrome has also been reported, but it is rarely seen.[6] The medications most commonly associated with the drug-induced form are all-trans retinoic acid and granulocyte colony-stimulating factor or granulocyte-macrophage colony-stimulating factor; others that have been reported are the sulfa drugs, tetracycline, minocycline, nonsteroidal anti-inflammatory drugs, carbamazepine, oral contraceptives, hydralazine, and clindamycin.[1] Alper et al reported a case of neutrophilic dermatosis that occurred as a result of exposure to a radiocontrast agent.[7] In addition to inflammatory bowel disease, Sweet syndrome has been associated with several autoimmune and other inflammatory conditions as underlying etiologies; they include systemic lupus erythematosus, dermatomyositis, rheumatoid arthritis, and Sjögren syndrome.

Clinically, patients present with an abrupt onset of multiple tender, erythematous plaques or nodules. The lesions may be vesicular, bullous, or targetoid, and they may present on the face, neck, chest, back, and extremities. In rare cases, oral or genital lesions are present. Other manifestations include ophthalmologic involvement (6 to 72% of cases), fever (65%), and arthralgias and myalgias (17%).[1] The differential diagnosis thus includes bullous pyoderma gangrenosum, erythema multiforme, urticarial vasculitis, rheumatoid neutrophilic dermatosis, and bacterial, mycobacterial, and deep fungal infections.

Su and Liu proposed a set of two major and four minor diagnostic criteria.[6] The two major findings include the presence of (i) tender, painful eruptions of plaque-like nodules and (ii) neutrophilic infiltrates in the dermis; both must be present to establish a diagnosis of Sweet syndrome. To complete the diagnosis, two minor findings must also be present; the four include (i) illness preceded by fever or infection, (ii) illness with fever, arthralgias, conjunctivitis, and an underlying malignancy, (iii) leukocytosis, and (iv) a response to steroids and not to antibiotics. Sweet syndrome rarely presents with oral manifestations. Our patient presented with both oral and palatal lesions and referred otalgia. Notani et al reported a rare case of Sweet syndrome with a palatal ulceration.[8] First-line treatment for Sweet syndrome is prednisone at 0.5 to 1.5 mg/kg/day tapered over 2 to 4 weeks. A corticosteroid injected into the lesions or applied topically may also be used. However, in this case, Dexamethasone 8mg was administered. In view of the possibility of head and neck involvement and the potential for an underlying paraneoplastic process, it is important that otolaryngologists be aware of the diagnosis of this rare syndrome.

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